Applicant: Parsa Kazemi-Estarjani et al. Serial No.: 09/639,207 : August 14, 2000

the claims:

lease amend the claims as follows:

(Canceled) Claims 1-25

TECH CHILLIAN SOOS 26. (Currently Amended) A transgenic D. melanogaster comprising a transgene containing a plurality of CAG's and at least one CAA sequence encoding a polyglutamine repeat sequence, wherein the repeat comprises at least 100 contiguous glutamine residues, and wherein the transgene produces polyglutamine toxicity in the transgenic D. melanogaster.

27-28 (Canceled)

- (Previously Amended) The D. melanogaster of claim 26, wherein the number of CAG's to CAA's is in ratio of between about 1:1 and 2:1.
- (Previously Amended) The D. melanogaster of claim 26, wherein the number of CAG's to CAA's is in ratio of between about 2:1 and 5:1.
- (Previously Amended) The D. melanogaster of claim 26, wherein the number of CAG's to CAA's is in ratio of between about 5:1 and 10:1.
- (Previously Amended) The D. melanogaster of claim 26, wherein the number of CAG's to CAA's is in ratio of between about 10:1 and 50:1.

Attorney's Docket No.:06618-686001

Applicant: Parsa Kazemi-Estarjani et al. Serial No.: 09/639,207

Filed : August 14, 2000

Page : 3

33. (Previously Amended) The *D. melanogaster* of claim 26, wherein expression of the polyglutamine sequence is conferred by a constitutive, regulatable or tissue specific expression control element.

- 34. (Previously Amended) The *D. melanogaster* of claim 33, wherein the tissue specific expression control element confers neural, retinal, muscle or mesoderm cell expression.
- 35. (Previously Amended) The *D. melanogaster* of claim 33, wherein the tissue specific expression control element comprises an Appl or rhodopsin 1 promoter or GLASS transcription factor element.

Claim 36 (Canceled)

- 37. (**Currently Amended**) The *D. melanogaster* of claim 26, wherein the polyglutamine sequence is between about 50 and 100 and 150 amino acids in length.
- 38. (Previously Amended) The *D. melanogaster* of claim 26, wherein the polyglutamine sequence is between about 100 and 200 amino acids in length.
- 39. (Currently Amended) The *D. melanogaster* of claim 26, wherein the polyglutamine sequence is between about $\frac{50}{200}$ and $\frac{250}{250}$ amino acids in length.
- 40. (Previously Amended) The *D. melanogaster* of claim 26, wherein the polyglutamine sequence further comprises a tag.

Applicant: Parsa Kazemi-Es arjani et al. Attorney's Jocket No.:06618-686001

'Serial No.:09/639,207 Filed : August 14, 2000

Page : 4

Claim 41 (Canceled)

- 42. (Previously Amended) The *D. melanogaster* of claim 26, wherein the Drosophila further comprises a marker sequence inserted into its genomic DNA, wherein the marker is located adjacent to a gene or inserted into a gene whose expression or activity increases or decreases polyglutamine toxicity in the animal, and wherein the marker sequence comprises an inducible upstream activating sequence, a minimal promoter sequence and 5' and 3' P transposon elements containing terminal inverted repeats.
- 43. (Previously Amended) The *D. melanogaster* of claim 42, wherein the marker sequence is near or inserted into a gene containing a J domain.
- 44. (Previously Amended) The *D. melanogaster* of claim 43, wherein the gene is HDJ1.
- 45. (Previously Amended) The *D. melanogaster* of claim 43, wherein the gene is TPR2.
- 46. (Previously Amended) The *D. melanogaster* of claim 43, wherein the marker sequence is near an MLF gene.
- 47-49 (Withdrawn)
- 50. (Previously Amended) A method of producing a transgenic *D.* melanogaster characterized by suppressed polyglutamine toxicity comprising:

Applicant: Parsa Kazemi-Estrjani et al. Attorney's Jocket No.:06618-686001

Serial No.:09/639,207 Filed : August 14, 2000

Page : 5

(a) transforming a *D. melanogaster* embryo or fertilized egg with a transgene comprising a plurality of CAA and CAG sequences encoding a polyglutamine sequence comprising at least 100 contiguous glutamine residues; and

(b) selecting a *D. melanogaster* that exhibits polyglutamine toxicity.

51-79 (Withdrawn)